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NEURO-FIBROMATOSIS OF THE TONGUE IN A CHILD, TOGETHER WITH A NOTE ON THE CLASSIFICATION OF INCOMPLETE AND ANOMALOUS CASES OF RECKLINGHAUSEN'S DISEASE.

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The patient, J. D—, a rather delicate-looking boy, aged 6 years when brought to the hospital, had a hard swelling below the tongue, which, according to the mother, had been almost certainly observed when the child was ten months old. The tumour in question (April, 1909) formed an oval projection (about 5 and 10 mm. in breadth

Fig. 1.—To show the position of the tumour under the tongue.

and length respectively) on the under surface of the tongue, situated along the right side of, and parallel to, the frænum linguæ, about half way between the tip of the tongue and the orifices of Wharton's ducts (see Fig. 1). The surface of the projecting tumour, which was apparently covered by healthy mucous membrane, was partly whitish and partly reddish in colour. No evidence of disease in the thoracic or abdominal viscera could be detected,

and the general health of the boy appeared satisfactory in spite of his somewhat delicate appearance.

The projecting portion of the tumour was kindly removed for me by Dr. Pfister; and Dr. J. C. G. Ledingham, to whom I am much indebted, kindly examined it microscopically. Sections (see Fig. 2) showed bundles of medullated nerve-fibres bound together by a close connective-tissue stroma. The tumour was evidently neuro-fibromatous, and a hard cord still remains to the right of the boy's frænum linguæ, doubtless representing part of the lingual branch of the fifth cranial nerve.

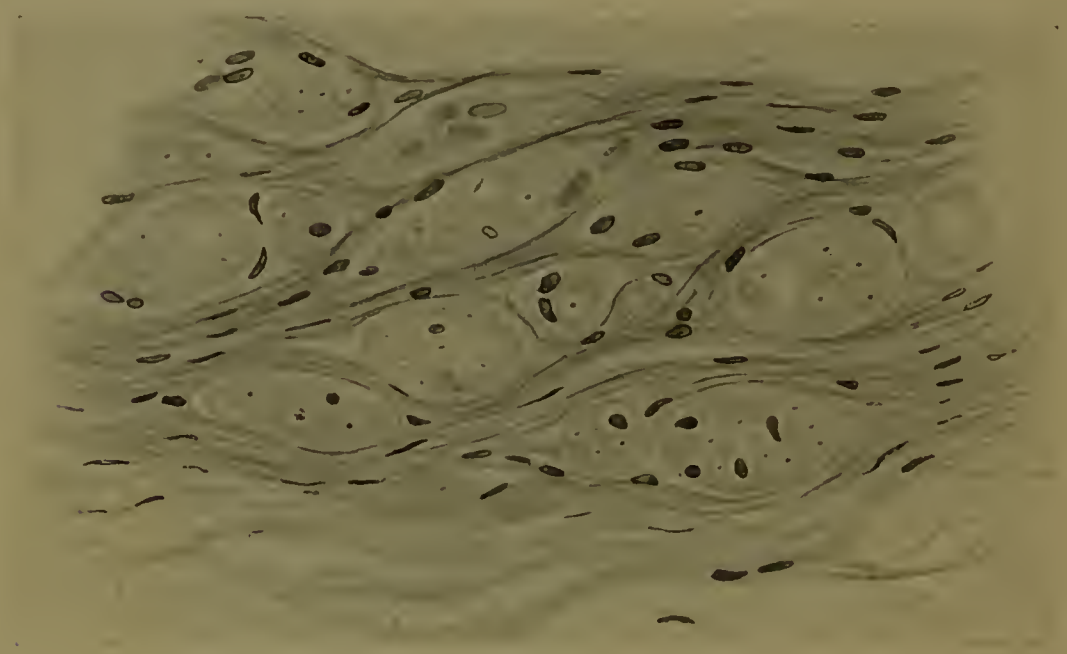

Fig. 2.—Microscopic section of a portion of the tumour, showing medullated nerve-fibres.

I regard the tumour as a mild form of plexiform neuroma involving one side of the tongue; in fact, I believe the condition to be a lesser degree of the "macroglossia (or rather hemi-macroglossia) neuro-fibromatosa" described by Abbott and Shattock in 1903,* and by Spencer and Shattock in 1907.† The case is, therefore, according to my classification, likewise to be considered as an (at least as yet) incomplete form of Recklinghausen's disease, and in this connection it is interesting to note that there is a somewhat irregularly shaped *café au lait* patch of cutaneous pigmentation, occupying an area of three or four square inches, on the upper part of the front of the

* Abbott and Shattock, 'Trans. Path. Soc., Lond.,' 1903, vol. liv, p. 231.

† Spencer and Shattock, 'Proc. Roy. Soc. Med.,' Pathological Section, 1908, vol. i, p. 8.

boy's left thigh. The mother thinks this patch of pigmentation was present from birth, in which case it might be termed "a superficial pigment-nævus." There is no abnormal pigmentation elsewhere on the body or limbs, nor are any neuro-fibromata detected except in the tongue. In this connection it may also be mentioned that a sister (aged 10½ years) of the patient has a small, flaccid ("empty"), molluscous-like tumour on the body (on the lower part of the sacral region to the left of the middle line), which is supposed to have been present at birth. She has no other tumours and no abnormal cutaneous pigmentation.

Though, strictly speaking, the term "Recklinghausen's disease" should be reserved for cases showing (1) obvious neuro-fibromata in connection with nerve-trunks, (2) molluscous tumours (molluscа fibrosa) of the skin, and (3) cutaneous pigmentation, yet incomplete forms occur in which one or even two of this triad of morbid features may be wanting. I would classify the anomalous or incomplete forms of Recklinghausen's disease as follows:

(1) Cases of plexiform neuroma and "elephantiasis nervorum" unaccompanied by multiple molluscous tumours of the skin, with or without cutaneous pigmentation. The least uncommon situations for these tumours are, perhaps, on the face and head. Into this group fall cases like the present one, and those of "macroglossia neuro-fibromatosa," already referred to.

(2) Cases of multiple molluscous tumours of the skin unaccompanied by any obvious neuro-fibromatosis of nerve-trunks, with or without decided cutaneous pigmentation.

(3) Cases of pigmentation of the skin not (or at least not as yet) accompanied by obvious neuro-fibromata of nerve-trunks or cutaneous neuro-fibromata (molluscous tumours). This class includes clinically all cases of cutaneous pigmentation of a kind similar to that met with in Recklinghausen's disease before the development of obvious neuro-fibromata,* though in such cases there may be neuro-fibromata present which cannot yet be detected by clinical examination. This class was not specially recognised, or given a special place, in Alexis Thomson's classical monograph, 'On Neuroma and Neuro-fibromatosis,' published at Edinburgh in 1900.

(4) Anomalous cases of neuro-fibromatosis, complicated by the co-existence of bony or epidermic (papillomatous) changes. Benaky† described an example of this class under the heading, "General

* *Cf.* F. P. Weber, "Cutaneous Pigmentation as an Incomplete Form of Recklinghausen's Disease," 'Brit. Journ. Derm.,' London, 1909, vol xxi, p. 49.

† 'Ann. de Derm. et de Syph.,' Paris, November, 1904, p. 977.

Neuro-fibromatosis, with Molluscum Pendulum of the Right Side of the Face." In his case there were deformities of the cranium, vertebræ, and tibia. In February, 1906, Mr. Lndford Cooper brought forward a case at the Ophthalmological Society, which he described as "neuromatous elephantiasis," in a girl, aged 11 years. In Cooper's case the outer and lower portions of the frontal bone and the squamous portion of the temporal bone were much more prominent on the affected than on the other side. In 1901 Sir Jonathan Hutchinson * described and figured the case of a woman with multiple molluscous tumours of the skin, whose right cheek was bulged by bony overgrowth; the right frontal bone was likewise thickened, and the right eye was pushed forward as if by bony growth behind it. At the Society for the Study of Disease in Children, in April, 1907, Dr. Fletcher Beach † showed a case described as one of Recklinghausen's disease, in a boy, aged 5 years and 10 months, who apparently also had bony thickening in the right temporal region. The most extreme example of this class was doubtless the famous "elephant man," whom many must have seen when he was at the London Hospital. Sir Frederick Treves,‡ in his description of this "elephant man," mentioned that the deformities of the osseous system were limited to the skull, right upper-extremity, and feet. "The proportions of the head were enormously increased, and its general outline was that of a hydrocephalic skull." There was exuberant papillomatous growth of some parts of his skin.

* 'Polyclinic,' London, July, 1901, p. 12.

† 'Reports of the Society for the Study of Disease in Children,' London, 1907, vol. vii, p. 167.

‡ 'Trans. Path. Soc., Lond.,' 1885, vol. xxxvi, p. 494.

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